WELCOME TO
AK LIVER DISEASE ECHO

ALASKA NATIVE TRIBAL HEALTH CONSORTIUM

NPAIHB
Indian Leadership for Indian Health

This project is supported by a grant from the Northwest Portland Area Indian Health Board and funding is provided from the HHS Secretary's Minority HIV/AIDS Fund.

PLEASE PUT IN THE CHAT BOX:

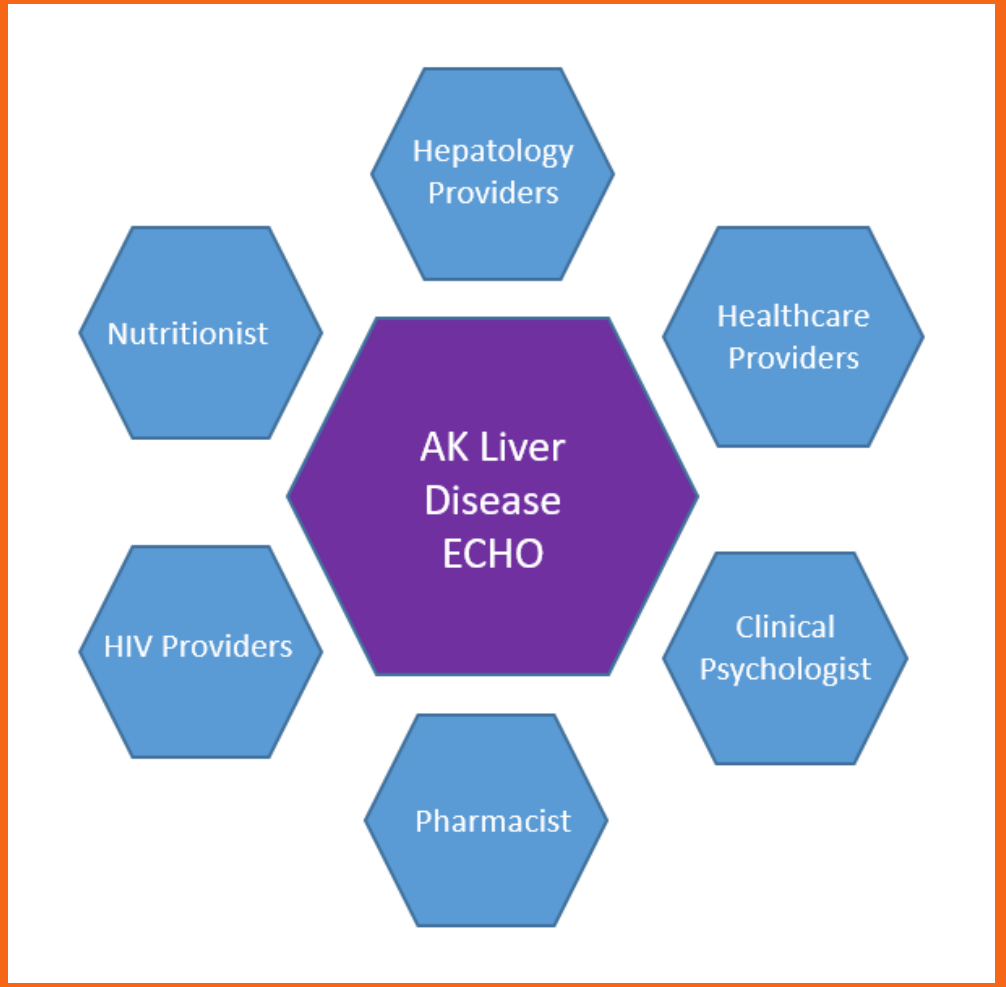
Your name
Where you are located
What brings you to the LD ECHO today

WHAT WE DO

- Didactic Presentations pertaining to ECHO topics
- We're accepting **case presentations and questions pertaining to:**
  - Elevated Liver Function Tests
  - Cirrhosis
  - Managing Complications of Decompensated Cirrhosis – Ascites, encephalopathy, esophageal varices
  - Alcohol-related liver disease, including Alcohol Hepatitis
  - Autoimmune liver disease – Autoimmune Hepatitis, Primary Biliary Cholangitis, Overlap
  - Nonalcoholic fatty liver disease/Nonalcoholic steatohepatitis
  - Hepatocellular carcinoma
- Provide Expert Panelists

CONSULTANT TEAM

- Brian McMahon, MD Hepatologist
- Youssef Barbour, MD Hepatologist
- Lisa Townshend, ANP Hepatology Provider
- Annette Hewitt, ANP Hepatology Provider
- Leah Besh, PA-C HIV/Hepatology Provider
- Anne Fleetwood, MS, RDN, NDN
- Brittany Keener, PharmD, MPH, BCPS
- Kena Desai, MD, Internal Medicine Specialist



Welcome to Alaska Liver Disease ECHO

Approved Provider Statements:

JOINTLY ACCREDITED PROVIDER™
INTERPROFESSIONAL CONTINUING EDUCATION

In support of improving patient care, Alaska Native Medical Center (ANMC) is jointly accredited by the Accreditation Council for Continuing Medical Education (ACCME), the Accreditation Council for Pharmacy Education (ACPE), and the American Nurses Credentialing Center (ANCC), to provide continuing education for the healthcare team.

Contact Hours:

ANMC designates this activity for a maximum of 12 contact hours, including 3 total pharmacotherapeutics contact hours, commensurate with participation.

Financial Disclosures:

Youssef Barbour, MD & Lisa Townshend-Bulson, APRN / faculty for this educational event, are primary investigators in an ANTHC sponsored hepatitis C study funded in part by Gilead Sciences. All of the relevant financial relationships listed have been mitigated.

Requirements for Successful Completion:

To receive CE credit please make sure you have actively engaged in the entire activity, your attendance is recorded by the facilitator, and complete the course evaluation form found here: https://forms.gle/R8vibUZgMbRcoScw9.

For more information contact
jlfielder@anthc.org or (907) 729-1387

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LAND ACKNOWLEDGEMENT

Out of great respect, we acknowledge the Creator of all living things and the original inhabitants upon whose traditional lands we respectively now gather or reside.

Let us, forever, remember the benefits and bounty of the vast resources that stem from these traditional lands, and thank the original inhabitants for their past and present stewardship of the resources (i.e., waters, plants, animals), and the spiritual practices which they cherish on the lands we now call home.

Primary Biliary Cholangitis

YOUSSEF BARBOUR M.D

Objectives

- PBC diagnosis
- PBC overlap syndromes
- PBC treatment

Pre-test

Which of the following is correct:

- 1- liver biopsy is needed to diagnose PBC
- 2- PBC affect intra and extra hepatic bile ducts
- 3- when alkaline phosphatase exceeds 1000 range it is recommended to start treatment using both Ursodiol and Obeticholic acid
- 4- Fatigue is the most common symptom of PBC

Introduction

Immune-mediated Cholangiopathies

- Immune-mediated cholangiopathies are chronic cholestatic disorders whose development is driven by auto- and allo-immunity. They include:
- **primary biliary cholangitis (PBC)**
- **primary sclerosing cholangitis (PSC)**
- **IgG4-related sclerosing cholangitis**
- **graft-versus-host disease**
- **hepatic allograft rejection**
- some studies also consider **biliary atresia** as an altered immunity-associated disease
- These diseases affect the biliary tree at different levels and for different extent, and they may also involve the peribiliary glands
- Immune-mediated cholangiopathies are characterized by an accumulation of activated auto- or allo-reactive **T lymphocytes** at the site of bile duct destruction, as the **regulatory T cell** is the main effector in the initiation of the process.

Main autoimmune biliary disease

Primary Biliary Cholangitis

- Destruction of small ***intrahepatic*** bile ducts

Primary Sclerosing Cholangitis

- multifocal biliary strictures and progression to end-stage liver disease. ***Intra- and extrahepatic*** bile ducts are primarily affected

Epidemiology, Etiology, Diagnosis

- PBC is mainly diagnosed in women (92% of patients), with a female:male ratio of about 10:1, and a mean age at presentation of 55 years.
- Population-based epidemiological studies across Europe, North America, Asia, and Australia have revealed an incidence of 0.9 to 5.8 per 100,000 people per year.
- The prevalence of the disease ranges from 1.9 to 40.2 per 100,000 people, and has also grown over time
- **Etiology** believed to be a combination of genetic and environmental trigger
- **Diagnosis**: chronic (at least 6 months) unexplained elevation in Alkaline Phosphatase, with positive AMA.. Liver biopsy is rarely needed.

Special cases

AMA positive, negative PBC

- Isolated positive AMA, with normal Alkaline phosphatase.
- AMA may be detectable in serum when patients are symptom free and liver tests are normal. Long term follow-up of 229 AMA-positive individuals for up to 7 years found that the 5-year incidence of PBC was **16%**.

AMA negative, positive PBC

- In a typical clinical picture of PBC, with chronic elevation in Alkaline phosphatase, about 5% of cases AMA can be negative (little higher in our population)
- PBC-specific ANA, including **sp100** and **gp210**, which are present in over 30% of PBC patients negative for AMA by indirect immunofluorescence. More recently, **anti kelch-like 12** and **anti-hexokinase 1** have been found in 35% and 22% of AMA-negative PBC patients, respectively, but these are not yet widely available.
- Liver biopsy is recommended if PBC specific Abs are negative

Clinical picture

PBC symptoms

- Fatigue
- pruritus (can be severe)
- Abdominal pain, about 17%, usually resolve spontaneously, and not related to disease stage

Associated diseases

- Sjogren's Syndrome
- Raynaud syndrome
- Hashimoto's Thyroiditis
- Osteoporosis
- CREST (calcinosis, Raynaud, esophageal dysfunction, sclerodactyly, and telangectasias)
- and systemic sclerosis

Biochemical tests in PBC

- In patients without cirrhosis:
- the degree of elevation in ALP is strongly related to the severity of ductopenia and inflammation
- the increase in aminotransferase activity and IgG levels mainly reflects the degree of periportal and lobular necrosis and inflammation
- and hyperbilirubinemia reflects the severity of ductopenia and biliary piecemeal necrosis.

Assessing biochemical response after 1 year f therapy

- **Rochester I** :ALP 2× ULN
- **Barcelona**: Reduction in ALP 40% from baseline or normalization of ALP
- **Paris I**: ALP 3× ULN; AST 2× ULN; and TB 1 mg/dL
- **Rotterdam**: TB 1× LLN
- **Toronto**: ALP 1.67× ULN
- **Paris II**: ALP 1.5× ULN; AST 1.5× ULN; and TB 1 mg/dL
- **Rochester II**: ALP 2× ULN
- **Global**: ALP 2× ULN

PBC overlap with AIH

- PBC and AIH can occur together, or one after the other.
- Some case are PBC dominant, and some are AIH dominant
- Diagnosis is based on biochemical pattern, autoantibody pattern, clinical manifestations.
- Liver biopsy may be indicated
- Treatment is toward the dominant disease

PBC therapy

PBC liver disease

- **UDCA** 13-15 mg/kg daily
- **OCA: Obeticholic acid** starting at 5 mg then titrate to 10 mg, daily or every other day depending on tolerance. (pruritus is the main side effect, and is contraindicated in decompensated liver cirrhosis)
- **Fibrates**, an off-label use for PBC

PBC pruritus

- Cholestyramine
- Rifampicin
- Opioid antagonists (naltrexone)
- Other treatment: Sertraline, Phenobarbital, antihistamines

Screening family members

- screening is usually recommended for female FDRs beginning at age 30.
- Screening is usually done by measuring the serum ALP level and, if it is elevated, assessing for AMA; this could be repeated at 5-year intervals if AMA-negative initially

Post test

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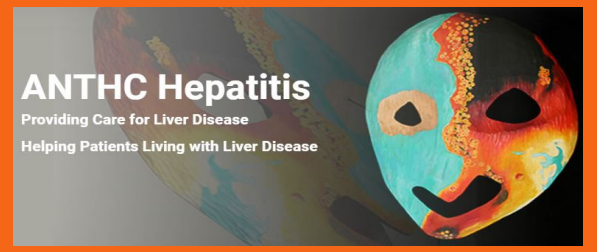
LIVER DISEASE ECHO SCHEDULE AT A GLANCE

- **July 21st – PBC/Overlap**
- August 18th – Most Common Liver Toxic Drugs
- September 15th – Trauma Informed Care and Liver Disease
- October 20th – Screening for Alcohol Use Disorder

ADDITIONAL LEARNING OPPORTUNITIES

- AK ID ECHO: HCV, HIV, PrEP, STIs
  - Second Tuesday of every month from 12:00-1:00PM Alaska Standard Time
  - August 9: HCV Reinfection vs Treatment Failure
  - 1CE/CME offered per session
  - anthc.org/ak-id-echo
- LiverConnect Webinar Program
  - Second Tuesday of every month 8:00-9:00AM Alaska Standard Time
  - August 9: Management of Complications of Cirrhosis
  - Full Hour didactic topics on Liver Disease and related topics 1CE/CME offered
  - anthc.org/what-we-do/clinical-and-research-services/hep/liverconnect/



HCV SIMPLIFIED TREATMENT TRAINING

- Receive the most current updates in patient care for screening and treatment of HCV.
- This training covers topics of HCV EPI, screening, confirmation of HCV, assessment of patient before treatment, HCV treatment, medication coverage, follow up and harm reduction.
- Continuing education credits available for enduring access credits.
- Presented by: Brian McMahon, MD, Leah Besh, PA-C, Lisa Townshend-Bulson, APRN, FNP-C and Annette Hewitt, APRN, FNP-C
- To complete the training, https://www.anthc.org/provider-resources/new-provider-webinars
- *For more information contact Marla at mjwehrli@anthc.org.*

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AK LIVER DISEASE ECHO -TEAM CONTACTS

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Thank you

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